Biliary Atresia Screening FAQ

Health Care Providers

1. What is biliary atresia?

Biliary atresia is a uniquely paediatric liver disease causing a progressive obliteration of the bile ducts, restricting bile flow from the liver to the intestine (cholestasis). It is the most common cause of liver transplant and liver related death in children.

2. Why are we screening for Biliary Atresia?

Screening can help identify babies with cholestasis earlier and potentially improve the chance of native liver survival. Babies with early-stage biliary atresia appear healthy and we know that in Ontario most babies with biliary atresia are currently not being diagnosed early enough to give them the best possible outcomes from their treatment.

3. How do families screen for biliary atresia?

Families compare their baby's stool to the pictures on the infant stool colour card (ISCC) during diaper changes. If the closest match is 1,2,3,4,5 or 6, they are to contact Newborn Screening Ontario using one of the several methods listed on the card so they can be guided through the next steps in the screening process.

4. What happens next, after a caregiver contacts NSO with concerns about pale stools?

- A team member from NSO will contact the caregiver within 1 business day to conduct a telephone assessment of the baby which will include questions about the baby's stool and other questions about the baby's health.
    - The team member may request that a photo of the stool be submitted as well.
- If the team member concludes that the stool is abnormally pale, they will facilitate an urgent remote consultation with a gastroenterologist at one of five paediatric academic health centres in Ontario.
- The GI specialist will then arrange for a total and direct/conjugated bilirubin to be done, will interpret the result, and will arrange next steps with the family.
- NSO will send a notification to the baby's primary care provider about the telephone assessment.

5. What are the signs and symptoms of biliary atresia?

- Babies with biliary atresia can appear healthy at first. Initially, they have prolonged jaundice lasting longer than 2 weeks of age and develop acholic (pale) stools, hepatomegaly, and dark urine.
- Babies with late-stage biliary atresia can go on to develop ascites, splenomegaly, and GI bleeding resulting from portal hypertension, along with failure to thrive, irreversible liver cirrhosis, and eventual death if surgery or liver transplant are not performed.

6. Why is the stool paler than normal?

Bile is a dark green to yellowish pigmented digestive enzyme that is made in the liver and flows into the intestine. It makes stool darker. In biliary atresia, the bile cannot get into the intestine resulting in stool that appears lighter in colour.

7. How long are families advised to screen for biliary atresia?

Families are advised to screen the stool using the ISCC for one month during diaper changes. Families of babies who are born before 37 weeks are advised to screen for one month past the actual due date.

8. What can I do to support biliary atresia screening for Ontario families?

You can ensure that families have an ISCC to screen at home. You can also offer education on BA screening as well as emphasize the importance of screening.

9. How can I get Infant Stool Colour Cards to distribute to families?

- If you are an existing NSO submitter, you can order Infant Stool Colour Cards from VWR International along with dried bloodspot collection cards as described here.
  www.newbornscreening.on.ca/supplies
- If you are not an existing NSO submitter, please contact us by email at nsoba@cheo.on.ca.

10. Does pale stool mean the baby has biliary atresia?

Stool screening with the infant stool colour card is not a diagnostic test. It tells us which babies have pale stools, possibly indicating cholestasis. There are several reasons why stool may be pale in colour and several causes of cholestasis, aside from biliary atresia. To make a diagnosis, other diagnostics such as blood tests and imaging of the liver are needed.

11. What is the treatment if a baby is found to have biliary atresia?

The treatment for biliary atresia is a surgical procedure called the Kasai portoenterostomy (KPE), where the flow of bile is re-established from the liver to the intestine. Despite receiving a KPE, some babies will still develop liver failure and will need to proceed to liver transplant. The earlier that this procedure can happen, the better the chance of native liver survival. Unless bile flow is established by surgical intervention through a KPE, children with biliary atresia rarely survive beyond 2 years of age without liver transplantation and mortality is inevitable in cases where no surgical repair or liver transplantation is done.